BEWARE OF SPHENOORBITAL MENINGIOMA

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ABSTRACT

Sphenoorbital meningioma is one of the tumors of the central nervous system with a high rate of recurrence in around three years. This case report deals with a late case of sphenoorbital meningioma. A female, 52 years old, came with a headache, the right eye stood out, and vision decreased dramatically from two to three years ago. Radiological imaging indicates the presence of a dextra sphenoid region meningioma with hyperostosis on the sphenoid wing that extends to the extra-conal. This case report shows a late case of sphenoorbital meningioma and recommends the need for early detection of sphenoorbital meningioma in order to improve the patient's prognosis.

Keywords: Early Detection; Female; Recurrence; Sphenoorbital

Received: Month Year, *Accepted: Month Year,* *Published: Month Year*

INTRODUCTION

Meningioma is one of the intracranial benign tumors that is commonly found with an incidence twice as common in women as in men (Saito et al., 2023). The incidence of meningioma is 12-15% of the intracranial tumor. Sphenoorbital meningioma is found in only 2-9% of all intracranial meningiomas (Masalha et al., 2021). Sphenoorbital meningioma is derived from the sphenoid wing with intraorbital extension and hyperostosis of the sphenoid bone (Atusufumi et al., 2019).

Sphenoorbital meningioma is a tumor that develops slowly and is characterized by the presence of proptosis or visual impairment (Baucher et al., 2024). The average age of patients at the time of diagnosis was 63 years, with details of 70%, 28%, and 3% each suffering from WHO meningioma grade I, II, and III. (Holleczek et al., 2019). The sphenoorbital meningioma itself is often recurrent. Recurrence incidence was 10.9% (26/239 patients), with an average recurrence time of 33.2 months (6-105 months) (Haddad et al., 2020). The risk factor for recurrence is the presence of invasion of the entire orbital, involvement of the orbital apex, superior orbital fissure, infratemporal fossa, and sphenoidal-ethmoidal sinuses, as well as World Health Organization grade II meningioma (Mariniello et al., 2022).

This case report shows a late case of meningioma in the sphenoorbital region that most likely will recur, which requires detection and caution against resurrence.

CASE

A female, Mrs. E, 52 years old, came to the hospital with a headache and the right eye protruding. The right protruding eye occurred with red eyes and blurred vision from two to three years ago. The patient also complained of nausea but didn't vomit. The patient went first to an ophthalmologist, who found that the patient's right-eye vision had decreased and suspected thre was a tumor. Then the patient

was reffered to a neurosurgeon for further examination. This patient had no history of oral contraceptives, regular use of corticosteroids and blood pressure medications, no radiation exposure, no family history of meningioma, or genetic syndrome.

Physical diagnose: GCS 15, blood pressure 131/89 mmHg, heart rate 99x/minute, respiratory rate 20x/minute, and temperature 36°C. The general physical examination showed the right eye can only see light (visual acuity 1/~), and the movement of the right eyeball is restricted. The patient weighs 70 kg with a height of 156 cm so the patient's body mass index based on Asia-Pacific is 28.76 kg/$m2$ (Obese I).

Head CT-scan with contrast obtained isodense lesions on the extra-axial dextra of the sphenoid region, relatively rounded shape, size 49x52x57 mm, relatively strict boundary, regular edge, pre-contrast density 44 HU and strong post-contrast 105 HU, visible hyperostosis on the dextra sphenoid wing, visible lesions extending to the extra-conal and pushing the dextra orbital structure into the anterior direction with the distance to the inter zygomatic line dextra 25 mm, sinistra 14 mm, directing the image of the meningioma on dextra sphenoid region accompanied by a picture of hyperostosis in the dextra sphenoid wing that extends to the extraconal that causing proptosis (Figure 1).

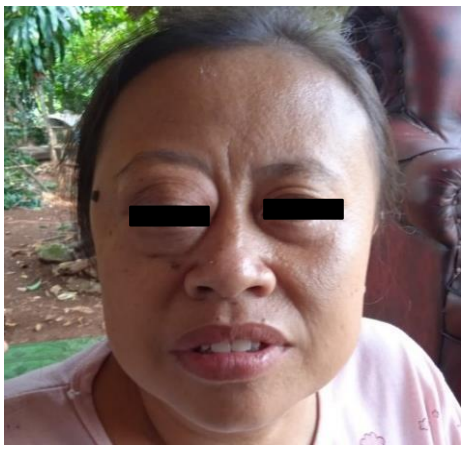

Figure 1. Preoperative clinical appearance from the right protruding eye.

A week after the radiological examination, the patient underwent craniotomy surgery to remove the tumor and install a subcutaneous drain. At the time of the operation, a sphenoorbital mass extending to the orbital roof was found with a soft consistency, dim color, and easily bleeding. A drill is used to make the orbital roof flat with no bone prosthetic.

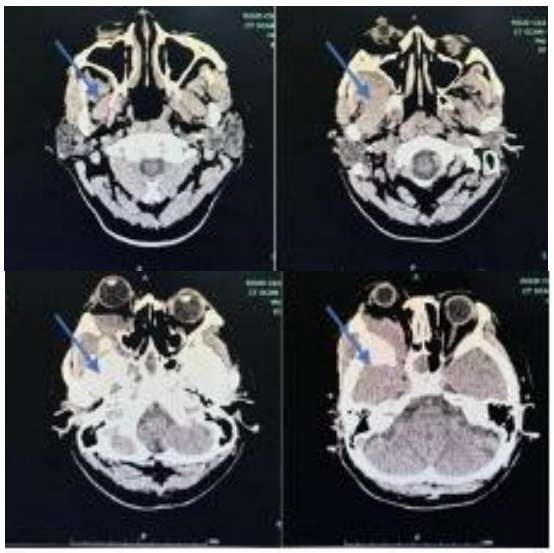

Figure 2. Head CT-scan with non-contrast.

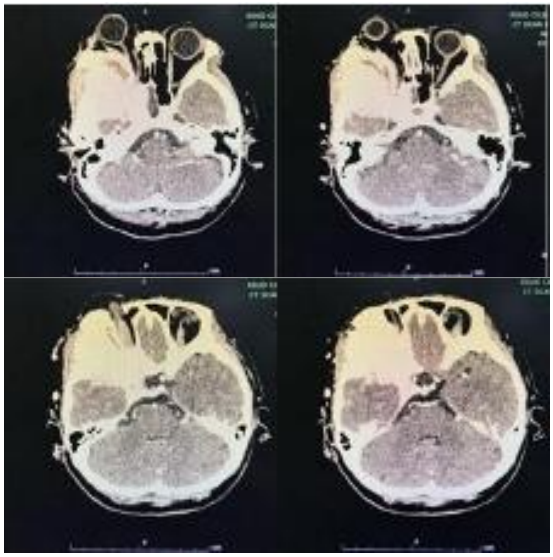

Figure 3. Head CT-scan with contrast showed isodense lesions on the extra-axial and hyperostosis on dextra sphenoid wing (pointed by blue thick line-arrow).

Microscopic findings found some mass consists of pieces of muscle tissue, bone trabecula and sequester. There was a mass of tumors that were whirling and solid. Round/ovular tumor cells, fine chromatin, and eosinophilic cytoplasm. Fibrotic stroma with congestive and hyperemic blood vessels. Diagnose: meningothelial meningioma (WHO Grade 1) (Figure 2 and 3).

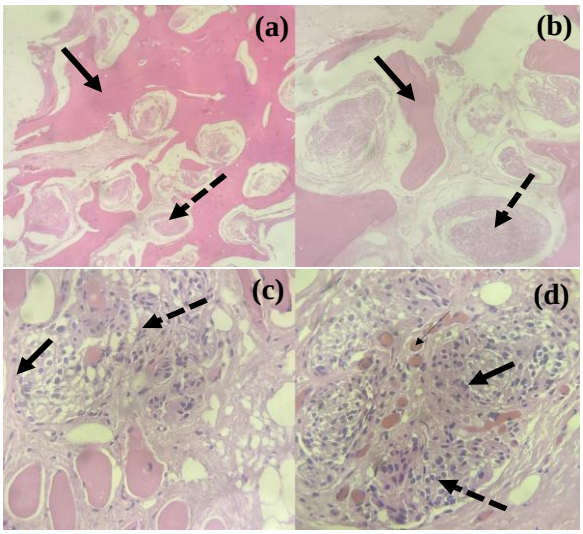

Figure 4. Microscopic appearance of meningioma meningothelial with 100x magnification for a and b, and 400x magnification, haematoxilin eosin staining for c and d).
(a) Thick-line arrow: bone trabeculam dotted-line arrow: a mass of tumors.
(b) Thick-line arrow: bone trabecula, dotted-line arrow: whirling and solid mass of tumors.
(c) Thick-line arrow: intranuclear pseudoinclusion, dotted-line arrow: eosinophilic cytoplasm.
(d) Thick-line arrow: syncytial cell, dotted-line arrow: uniform round cell, thin-line arrow: congestive and hyperemic blood vassels.

DISCUSSION

Sphenoorbital meningioma is a primary enplaque tumor on the sphenoid wing that attacks the surrounding anatomical structure. Patients are usually women in their 50s, come with progressive, unilateral, non-pulsatile proptosis, and often have complaints of cosmetic deformities and optic verve damage (Wierzhowska et al., 2022; Baucher et al., 2021). Patients with sphenoorbital meningioma usually experience trias of clinical symptoms including proptosis, reduced vision, and ocular paresis (Saito et al., 2023). As in this 52-year-old woman patient, she was experienced all those trias, and her symptoms were progressive over the years. The patient ended up getting her medication after she had a headache.

Using corticosteroids, using blood pressure medications, having a high body mass index, and being a woman are some risk factors linked to meningioma (Cerhan et al., 2019). As in this patient, she is a woman and has a high body mass index. Environmental factors like obesity, exposure to ionizing radiation, radiators therapy, exogenous hormonal factors such as hormone replacement therapy can increase the risk of meningioma (Ogasawara et al., 2021). Due to the patient's classification as Obese I, obesity is most certainly the risk factor for the patient. Obesity is associated with higher levels of IGF-1 (insulin growth factor-1), which is known to suppress apoptosis and accelerate tumor formation (Jin et al., 2023). A greater expression level of IGF signaling system components has also been seen in meningiomas, suggesting a possible role for these proteins in the tumor progression (Takahashi et al., 2019).

The definitive treatment is to perform a resection surgery. To achieve complete resection and regression of apoptosis, it is important to drill the lateral and/or superior orbital wall sufficiently and carefully, remove any soft tissue components inside the orbit, and remove any accessible hyperostotic bone (Fathalla et al., 2020). Extension of the hyperostosis into the inferior orbital walls, medial orbital walls, or infra-temporal fossa are factors that make entire excision challenging. Furthermore, is it known that achieving complete excision might be difficult when a tumor adheres to the orbital muscles or extends into the cavernous sinus (Elborady and Nazim, 2021). A s in this patient, seen in preoperative imaging with a

condition of hyperostosis on the dextra sphenoid wing that already extended to the extraconal area.

This patient's microscope findings were meningothelial meningioma. It is the most common variation in WHO classification grade 1, with a lobulated-circular architecture, often contains meningothelial whorls. Syncytial cells (some cells that merge into two or more) with indistinct cell membranes, eosinophilic cytoplasm, uniform round nucleus, intranuclear pseudoinclusion common occur, and may have psammoma bodies (Cai, 2021).

Only a small percentage of meningiomas are aggressive, such as those the World Health Organization (WHO) classifies as grade II and III; the majority are grade I meningiomas, which are occasionally referred to as "benign." Even yet, prior research has shown that recurrence rates for WHO grade I meningiomas can reach 47% with long-term follow-up. Prognostic indicators for WHO grade I meningioma recurrence include subtotal resection, posterior fossa location, nuclear atypia, and increased MIB-1 index (Peyre et al., 2017; Haddad et al., 2020). In patients with sphenoorbital meningioma, the maximal safe resection continues to be the most significant prognostic factor linked for reduced recurrence rates (Masalha et al., 2021). As in this patient that classified as meningothelial meningioma (WHO grade 1), but the resection was sub-total due to the location of the tumor on the dextra sphenoid wing that extends to the extra-conal. Therefore, the patient underwent maximal resection with the goal of performing decompression of the optic nerve and excision of the hyperostosis bone in order to lessen the proptosis condition (Figure 4).

The tumor gradually grows again during the five years following surgery. It is crucial to do long-term monitoring to identify early-stage recurrence. For recurring tumors, further surgery to decompress the brain structures should be contemplated; stereotactic surgery may be a viable option for malignant or recurrent instances (Nagahama et al., 2019).

A multidisciplinary strategy with a skull base team is necessary to maximize patient outcomes. Radiologists, ophthalmologists, otorhinolaryngologists, maxillofacial surgeons, and neurosurgeons make up this team. Through their combined experience, the team members are able to promote enhanced surgical resection and clinical outcomes, as well as early diagnosis of optic nerve impairment and evidence-based care both preoperatively and postoperatively (Almaghrabi et al., 2023; Agosti et al., 2023).

Further research is needed to explore the pathogenic mechanisms of meningioma tumors so that effective early detection methods and therapeutic protocols can be found. Despite this, it is challenging to discover meningioma early in all patients due to the challenge of managing the disease's underlying etiology. Therefore, consensus and approaches are needed both genomically and epigenomically as forms of early detection (Himič et al., 2023; Zhao et al., 2020). Meningioma progressivity may now be determined postoperatively using genomic and epigenomic detection, which includes chromosomal loss or gene mutation (Deng et al., 2022; Rawanduzy et al., 2023). Therefore, more study is required to enable early meningioma illness identification

The weakness from this case report doesn't provide much information about the patient's lifestyle or risk factor. In addition, although this report briefly mentions recurrence, it does not provide enough information about the patient's prognosis or the likelihood of recurrence following treatment. This leaves a gap in understanding the potential challenges in managing the condition in the long term.

Despite being uncommon, Sphenoorbital meningiomas are important because they frequently return within three years of initial therapy. Given that symptoms including headaches, visual abnormalities, and eye protrusion can indicate the presence of this tumor, this instance emphasizes the

significance of early identification. Screening is essential for both diagnosis and assessing the tumor's extent, especially MRI with contrast. Surgical resection is usually the method of management; if resection is not feasible, adjuvant radiation therapy may be required. Long-term follow-up with routine imaging is necessary to check for any indications of tumor regrowth because of the high recurrence rate. A multidisciplinary approach is essential for successful therapy, and prompt diagnosis and action can enhance the patient's prognosis (Nogueras and Jagelman, 1993; Martucci et al., 2023).

CONCLUSION

This case report shows a patient with sphenoorbital meningioma who came late to seek treatment. As a serult of its late arrival, the tumor has spread and invaded many other parts so that resection can only be performed sub-total, where subtotal resection is one of the determining factors for recurrence in patients with a rate of recurrence in WHO Grade I meningioma can reach 47% with long-term observation. So, it is learned that cases of sphenoorbital meningoma should be cautious, as often patients show no clinical symptoms at the beginning of the course of the disease. Therefore, early diagnosis is necessary so that proper treatment can be carried out promptly in order to improve the prognosis and quality of the patient's daily life.

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